346

Serum levels and prescribed dosages of vitamin A and E in patients with cystic fibrosis

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The **aim** of the study was to evaluate the serum levels of vitamin A and E, and compare those to prescribed doses and to normal serum levels in comparison to the recommendations given in the consensus report (1)
Material: One hundred-twelve consecutive patients, (males 64) attending the annual check-up at the West Swedish CF Centre in 2003 were included. Patients diagnosed less than a year before the annual review, pregnant, transplanted or patients waiting for transplantation were excluded. Ninety-two (82%) were pancreatic insufficient (PI). Mean age were 20.4± 12.0 years; 48% were ≥18 years.
Method: Serum levels of alpha-tocopherol and retinol were measured in the morning after one night fast. Mean±SD are given.
Results: S-retinol were 2.2±0.6µmol/l. Patients with PI had significantly lower s-retinol than patients with PS (p=0.03), without difference between adults and children. All but one patient had s-retinol levels within the reference range. There was no correlation between s-retinol levels and $FEV_{1.0}$ or age Prescribed amount of vitamin A were 4480 ± 2528 IU (range 2400-15900 IU) in patients with PI and PS. S-alpha-tocopherol was 26.2 ± 11.1µmol/l. Seven patients (all PI) had serum levels below the reference range. There was no correlation between s-α-tocopherol and $FEV_{1.0}$, PI/PS or age Prescribed amount of vitamin E was 260 ± 138 IU in the PI group and 81 ± 94 IU in the PS group.
Conclusion: The supplementation of vitamin A and E were within the recommendations and most patients had serum concentrations within the reference range. Low adherence could be one explanation to low serum levels of tocopherol in some patients despite the prescribed E-vitamin dosage.
1. Sinaasappel M et al. J Cyst Fibr 2002;1:51-75

347

An audit of additional vitamin D supplementation on vitamin D levels in children with cystic fibrosis

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Introduction There is increasing evidence that vitamin D deficiency and seasonal variability in plasma levels may predispose to the development of osteoporosis. Vitamin D is usually given in combination with vitamin A. Increasing the dose may lead to vitamin A toxicity and a separate vitamin D preparation may be required. We report the change in vitamin D levels over 4 years following the implementation of a vitamin D supplementation protocol.
Method 104 children, median age 7.3yrs (0.3-13.2) had plasma 25-hydroxy vitamin D levels measured in 2000. All were receiving 10-40 µg vitamin D combined with vitamin A. If the vitamin D level was below 30 ng/ml patients received a review of vitamin and pancreatic enzyme dosing and adherence. Additional supplementation with a preparation containing calcium and vitamin D (10-30 µg/d) was given as appropriate. Levels were reassessed in 2004.
Results The mean (SD) vitamin D levels for 2000 and 2004 were 19.5 ng/ml (7.5) and 25.3 ng/ml (7.5) respectively (p<0.001). The proportion of patients above 20 ng/ml and 30 ng/ml increased from 45% to 71% and 8% to 19% respectively. The percentage of patients in each season within a range of serum vitamin D values is shown below:

Season	Jan – Mar		Apr – Jun		July – Sept		Oct - Dec	
ng/ml	2000	2004	2000	2004	2000	2004	2000	2004
< 10	38	0	5	0	0	0	4	11
10 - 20	62	29	51	0	37	6	46	24
20 – 30	0	65	35	65	48	61	46	61
> 30	0	6	8	35	15	33	4	5

Discussion These results confirm a seasonal variation in vitamin D levels and demonstrate that a protocol aimed at increasing levels has resulted in a significant improvement. However, 94.5% of children in the winter months still had levels less than 30 ng/ml. This suggests that some patients need more vitamin D than is currently recommended in nutritional guidelines.

348

Vitamin D status in children with Cystic Fibrosis

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Background: Bone disease and low bone mineral density are recognised as long term complications in the adolescent and adult Cystic Fibrosis (CF) population. Sub-optimal levels of serum 25-OH Vitamin D (25-OHD) have been reported in the CF literature and may contribute to poor bone mineral density. Levels <25nmol/l are classed as deficient, however recent CF literature has suggested that serum 25-OHD levels should be maintained at 75 nmol/l-150 nmol/l to improve bone accretion. In our clinic all pancreatic insufficient children are prescribed a daily Vitamin D supplement of either 10 µg or 15 µg for infants and individuals > 1 year of age respectively. We aimed to determine the prevalence of Vitamin D deficiency in our CF population. **Methods**: The serum 25-OHD level of children attending the CF annual review were measured. Data were analysed according to age (0-5 years, 5-10 years and 10-16 years) and whether the serum 25-OHD levels were measured in winter, spring, summer or autumn months. **Results**: 187 children (aged 0-16 years) were studied. The mean (±SD) serum 25-OHD level was 48.30 (18.03) nmol/l, range (21-125 nmol/l). Three percent had levels less than 25 nmol/l and 6% had levels > 75nmol/l. The mean serum 25-OHD in the summer months, 66.41(17.80) nmol/l, was significantly higher (p<0.001) than those measured during the rest of the year, winter 42.80 (14.90) nmol/l, spring 40.60 (12.69) nmol/l) and autumn 51.00 (16.54) nmol/l. This effect was independent of age. **Conclusion**: The majority of children were found to have 25-OHD levels > 25 nmol/l but < 75 nmol/l. To maintain serum 25-OHD levels consistently > 75 nmol/l our current Vitamin D supplementation regimen may not be adequate. Further research regarding optimal 25-OHD levels and adequate Vitamin D dosage to support this is needed.

349

Somatic development of CF children diagnosed in neonatal screening

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The **aim** of our study was to assess the nutritional status of children diagnosed in neonatal screening from 1999-2003.
Among 71 children diagnosed, 57 (27 girls, 30 boys) born in 1999-2003 are followed - up in the Warsaw CF Center.
The children were subdivided into 2 groups:
1. Pilot group-11 children born in 1999. Mean age of diagnosis was 15 weeks.
2. Main group –46 children born in 2000-2003. Mean age of CF diagnosis was 6 weeks.
All patients had regular weight and height measurements.
49 newborns were full term with mean birth weight (kg) female 3,161 (SD 0,44) and male 3,46 (SD 0,45)
8 premature babies with mean birth weight (kg) female 2,63 (SD 0,35) , male 2,19 (SD 0,6)
In 55 children PI was diagnosed. They were given pancreatic enzymes.
Only 2 children from the main group were PS (Δ F508/3849+10kbCT, Δ F508/?) with normal somatic development and normal elastase -1 in stool.
Somatic development was normal in 63% of children from the main group and 46% of the pilot group; dysharmonic – in 22% of the main group and 35 % of the pilot group; abnormal in 15 % of the main group and 19% of the pilot group.
The biggest changes in nutritional status were observed in the first year of life, especially in the pilot group. Subsequently nutritional status was gradually improving. At the end of the 4th year of life 100% of children from the main group had normal nutritional status compared to 80% from the pilot group.
Conclusions:
1. The mean birth weight of CF children from neonatal screening was not lower than mean birth weight of Polish healthy newborns.
2. Most infants presented clinical symptoms of pancreatic insufficiency
Early introduction of complex therapy including nutritional treatment with pancreatic enzymes and electrolytes supplements prevents severe malnutrition.